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Myasthenia Gravis
Reference Summary

Myasthenia gravis is a disease that affects the muscles, causing them to become weak. Medication is available to treat myasthenia gravis. However, medical emergencies can arise if the disease affects the breathing muscles. This reference summary will help you understand what myasthenia gravis is and how it can be treated. The causes and symptoms of this disease are reviewed also.

Myasthenia Gravis

Myasthenia gravis is a disease that causes weakness in the voluntary muscles of the body. This muscle weakness usually occurs during periods of activity and improves after periods of rest.

Certain muscles, such as the ones controlling eye movement, facial expression, chewing, talking, and swallowing are often involved in myasthenia gravis.

Myasthenia gravis may also affect the muscles that control neck and limb movement and breathing. *Myasthenia gravis* means "grave muscle weakness." However, due to recent advances in medicine, most cases of myasthenia gravis are not as grave, or serious, as the meaning implies.

Myasthenia gravis is not contagious or fatal and does not affect life expectancy. Myasthenia gravis affects all ethnic groups and both genders. It can occur at any age, however, it most commonly affects women under the age of 40 and men over the age of 60.

Causes

In order to understand the causes of myasthenia gravis, it is important to know how the nerves and muscles work together to make movement. It is also necessary to understand the body's immune system. The body has 2 kinds of muscles: voluntary and involuntary. We directly control voluntary muscles to make the body walk, run, write, lift, smile, and chew.

Involuntary muscles work on their own. These muscles are found in the stomach, intestines, colon, and blood vessels.

The brain controls voluntary muscles by sending messages through the nerves. Messages go from the brain to the spinal cord, then through the peripheral nerves to the muscles. Nerves are made of cells called neurons. A neuron has a long fiber called an axon that electrical impulses travel through.

When messages from the brain arrive at the end of a nerve, they are transmitted to the muscle. The space between a nerve ending and a muscle is called a neuromuscular junction.

As messages arrive to the neuromuscular junction from the brain in the form of electrical waves, they release, in that junction, special chemicals called neurotransmitters

When acetylcholine reaches the muscle side of a neuromuscular junction, it locks into special *keyholes*, causing the muscle to contract, these keyholes are known as receptors. If anything interferes with the steps needed to transmit brain messages across a neuromuscular junction, the nerves will not be able to control muscle contractions. Myasthenia gravis occurs when normal communication between nerves and muscles is interrupted at the neuromuscular junction. The interruption is due to antibodies that prevent acetylcholine from reaching the muscles by destroying or blocking the receptors. Antibodies are substances made by the body's immune system to fight germs and infections.

The thymus, a small gland located under the breastbone, is very important in the development of the immune system early in life. In adults with myasthenia gravis, the thymus gland is abnormal.

Some patients with myasthenia gravis develop abnormal growths or tumors in the thymus gland. These tumors are usually benign and non-cancerous. Although the relationship between the thymus glad and myasthenia gravis is not clear, scientists think the thymus gland may be responsible for the immune system attacking acetylcholine receptors.

Symptoms

Myasthenia gravis may affect any voluntary muscle. However, the muscles that are most commonly affected are those that control the eye and eyelid movement, facial expression, and swallowing.

Myasthenia gravis may happen suddenly. In most cases, the first noticeable symptom is weakness of the eye muscles that causes double vision. In some cases, difficulty swallowing and slurred speech may be the first signs.

The degree of muscle weakness involved in myasthenia gravis varies greatly among patients. Symptoms may include:

- unstable or waddling walk
- weakness in arms, hands, fingers, legs, and neck
- a change in facial expression difficulty swallowing
- shortness of breath
- impaired speech

A *myasthenic crisis* occurs when weakness affects the muscles that control breathing. This creates a medical emergency whereby the patient needs a respirator in order to breathe.

In patients whose respiratory muscles are weak, myasthenic crises may be triggered by infection, fever, side effects of medication, or emotional stress.

Diagnosis

Since weakness is a common symptom of many disorders, the diagnosis for myasthenia gravis is often overlooked in patients who experience only mild weakness or weakness in only a few muscles. To diagnose myasthenia gravis, a medical history, physical exam, and neurological exam must be performed. Myasthenia gravis is suspected if

- eye movements are impaired
- muscles are weak but the patient can still feel things normally.

Several tests are available to confirm the diagnosis of myasthenia gravis. A blood test can detect high levels of the antibodies responsible for attacking the acetylcholine receptors.

Another test is called the *edrophonium test*. If a myasthenia gravis patient with eye weakness is given edrophonium chloride, muscle weakness will be relieved within a few minutes.

Other tests that help diagnose myasthenia gravis include electrodiagnostic examination where the function of the nerves and muscles are studied.

CAT scans and MRI may be used to identify an abnormal thymus gland.

A special examination called pulmonary function testing, or PFT, can be done to measure breathing strength. It also helps predict whether respiration may fail and lead to a myasthenic crisis.

Treatment

Myasthenia gravis can be controlled with medication. Some medications help improve neuromuscular transmission and increase muscle strength. These medications are known as anticholinesterase agents such as neostigmine and pyridostigmine. They work by slowing down the breakdown of acetylcholine in the neuromuscular junction. The acetylcholine therefore has more time and increased chance to stimulate the muscle.

They do have some side effects and some time taking too much of these medications can actually lead to weakness and symptoms similar to myasthenia gravis. This is why it is important to take the medication as directed and to let your doctor know if there is any increase in weakness.

Other medications limit the production of antibodies. These medications can have major side effects. These are known as immunosuppressive drugs they include prednisone, cyclosporine, and azathioprine

For about 50% of myasthenia gravis patients, surgery can be done to remove the thymus gland, when it is abnormal. This surgical procedure, which is called thymectomy, may help improve the control of myasthenia gravis.

It is usually recommended for patients younger than 60 years of age. Older patients do not tend to benefit as much for a thymectomy. For some patients, removing the antibodies from the blood can be helpful. This procedure is called *plasmapheresis*.

Another treatment consists in giving the patient high-dose intravenous immune globulin, which temporarily modifies the immune system and provides the body with normal antibodies from donated blood. This is known as IVIG

The doctor determines which treatment option is best for each patient. This depends on several factors including

- how severe the weakness is
- which muscles are affected

- the patient's age and medical condition

Summary

Myasthenia gravis is a disease of the neuromuscular junction. It can lead to muscle weakness and possibly respiratory problems if not treated. Treatment for myasthenia gravis is available and very effective in controlling the disease. Most patients with myasthenia gravis can expect to live healthy normal lives with close to normal activity levels!